



Case report

A case of mycetoma-like chromoblastomycosis in Qatar

Gawahir A. Ali^a, Wael Goravey^a, Saad J. Taj-Aldeen^b, Mahir Petkar^b, Issam Al-Bozom^b, Hamad Abdel Hadi^{a,*}

^a Department of Infectious Diseases, Qatar

^b Department of Laboratory Medicine and Pathology Hamad Medical Corporation, Doha, Qatar



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ABSTRACT

Chromoblastomycosis is one of the neglected tropical mycoses associated with chronic infection of the skin and subcutaneous tissues. We report a case of 49-year-old patient originally from India presented with a mycetoma-like lesion in his right foot which was slowly progressing over three-year period. The diagnosis of chromoblastomycosis was confirmed following surgical excision and identification of the unique histological pathology of muriform bodies. The patient was subsequently treated with a prolonged course of oral itraconazole with a favorable outcome. The clinical presentations, assessment and management of the disease are outlined.

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Introduction

Chromoblastomycosis (CBM) is a chronic granulomatous mycosis of cutaneous and subcutaneous tissues caused by melanized dematiaceous fungi acquired usually by inoculation of the skin with the pigmented fungi as part of occupational or environmental hazards [1]. The most prevalent etiological agents are *Fonsecaea pedrosoi*, *Phialophora verrucosa* and *Cladophialophora carrionii* which are frequently involved in the majority of cases [2]. The disease has worldwide prevalence with predominant cases reported from humid tropical and subtropical areas such as regions in South America and the Indian subcontinent. The disease usually leads to chronic slow progressing infection with subsequent destruction of surrounding tissues occasionally complicated by secondary bacterial infections or malignant transformation [3]. The characteristic dark-colored, thick-walled, septate sclerotic cells coined copper pennies muriform cells and Medlar bodies are frequently observed, which are the suggestive histopathological criterion for the diagnosis [3–5]. The clinical presentation and diagnosis are usually delayed because of the slow progression of the disease with minimal presenting symptoms. Prolonged antifungal treatment in combination with appropriate surgical interventions such as excision, cryotherapy or thermotherapy is the mainstay of management with varying degree of success depending on lesions site, size and presentation [5,6].

In the present report we describe a case of chromoblastomycosis, which was successfully treated with surgical intervention followed by antifungal therapy.

Case description

A 49-year Indian male with no significant past medical history or chronic diseases presented to Hamad General Hospital, Doha, Qatar with a three years history of non-painful nodular swelling at the dorsum of the right foot (Figure 1). The patient recalled a history of trauma to the same area when previously farming in rural India. Over the 3 years period, there was gradual increase in the size of the swelling up to his presentation.

The initial medical assessment was of complicated cutaneous ganglion since clinical examination did not reveal any significant physical signs apart from right first web space non-tender nodular swelling measuring about 3 cm in diameter with minute sinuses. Serous discharges were expressed following deep manipulation of the swelling. Blood tests including complete blood counts, renal profile, liver function tests, blood glucose, glycosylated hemoglobin and inflammatory markers were unremarkable. Microbiological assessment of the fluid obtained from the lesion failed to isolate any pathogens.

Magnetic Resonance Imaging of the right foot (Figure 1) demonstrated a suspicious lesion with irregular multi-lobulated soft tissue mass overlying the distal first metatarsal bone accompanied with surrounding bony erosions. The radiological assessment was in favor of a chronic mycetoma-like infection rather than neoplasm. Following initial assessment, tourniquet assisted surgical excision and debridement of the surrounding tissues was performed

* Corresponding author at: Communicable Diseases Centre, Hamad Medical Corporation, Doha, Qatar.

E-mail address: habelhadi@hamad.qa (H.A. Hadi).

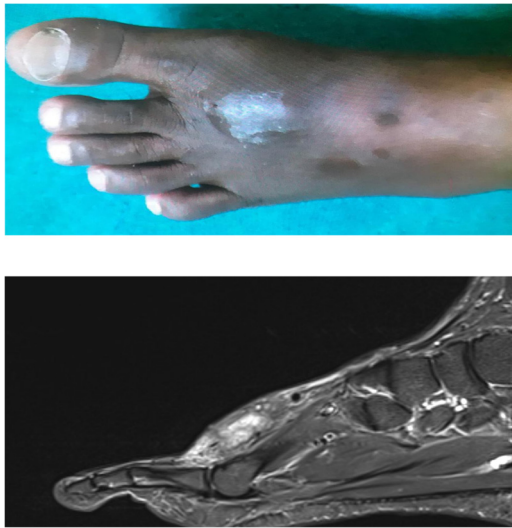


Fig. 1. Nodular swelling at the dorsum of the right foot overlying the first and second metatarsal bones. There are skin discoloration and desquamation from the underlying enlarging lesion.

MRI images demonstrated multi-lobulated soft tissue lesion overlying the distal part of the first metatarsal bone eroding its base suggestive of mycetoma.

in order to assess the underlying pathology as well as provide curative therapeutic measures.

Fungal and bacterial cultures of the obtained tissues failed to grow any pathogen while histopathological examination revealed necrotizing granulomatous inflammation, surrounded by numerous brown pigmented fungal organisms, arranged in the form of solid clusters of thick-walled fungal cells, displaying features compatible with chromoblastomycosis (Figure 2). These organisms were seen amid dense acute inflammation, forming abscesses.

Reviewing the history and presentation together with the histological diagnosis which was consistent with chronic fungal infection; the patient was started on a prolonged course Itraconazole 400 mg per day with favorable clinical response and no disease progression following one year of intervention.

Discussion

Chromoblastomycosis is a chronic granulomatous mycosis of cutaneous and subcutaneous tissues common in tropical and subtropical areas caused by ubiquitous melanized pigmented dematiaceous fungi reflected in its name [7]. The disease was historically described in Brazil towards the beginning of the 20th century and obtained multiple synonymous names till an international consensus in 1992 settled for the current disease classification of CBM [6].

The main mode of transmission is through cutaneous inoculation either through penetrating of the intact skin or contamination of superficial wounds. Bare feet workers or those being in direct contact with contaminated environmental samples containing melanized fungi are at significant risk.

Despite the ubiquitous nature of the different causative fungi in the environment, infection with CMB remains low with no accurate incidence figures; the highest endemic areas with reported cases are Madagascar in Africa and Brazil and Venezuela in South America. Infection is more prevalent in middle age groups between 30–50 years particularly those from agricultural background with male preponderance of almost 80% of cases [4,6]. Different susceptibility mechanisms have been implicated for the chronic fungal infection including protective nature of female sex hormones, genetical susceptibility as well as a defective immune mechanism to identify

the fungus. Conversely, immune suppression has also been implicated with the more aggressive form of the disease [6].

The absence or paucity of symptoms because of indolent progression are the main reason for delayed presentation ranging from weeks to years [2,3,6]. This could be slowing growing non tender lesions particularly at the foot or progressive plaques masquerading as common fungal dermatological diseases. Classification of clinical presentation of CBM has five distinctive forms of the slowly progressive lesions: nodular, tumor-like, verrucous, plaque and cicatricial depending on size, site, and location as well type of fungi [6]. Our case presented with slowly progressive nodular lesion which originates from deeper tissues but resembled a tumor during clinical assessment. Assessment particularly with deep seated infections entails radiological investigations particularly CT or MRI which are not only helpful in delineating soft tissues and bone involvement but also differentiating it from deeper neoplastic growth. The diagnosis is usually confirmed upon microscopy or histopathological examination in addition to appropriate fungal cultures to identify the organism for its characteristic pathology.

Common histopathological findings are epidermal hyperparakeratosis, pseudoepitheliomatous hyperplasia, intra-corneal micro abscesses and granuloma formation while the hallmark of CBM disease is the presence of dark colored and thick-walled cells coined muriform cells with sclerotic component which are the typical features of the chronic fungal infection [6].

The management of CBM is challenging since cases are either endemic or sporadic with absent wide supporting evidence and uniform guidelines. Cases have a wide presentation spectrum with high tendency of relapses following initial treatment success. The best management approach consists of a combination of a prolonged antifungal treatment particularly an azole agents, preferably itraconazole at doses of 400 mg per day or higher if tolerated, posaconazole at doses of 800 mg per day and/or terbinafine as they appear to have high in vitro activity against the etiological agents of CBM [6]. Frequently this is combined with physical treatment like excisional surgery, cryotherapy or thermotherapy according to the spectrum of presentation. The exact choice and success of different management modalities depends on the location involved, lesions extend and size as well as fungal species of CBM with variable cure rates ranging from 15% to 80% since some species like *Fonsecaea pedrosoi* is less responsive to antifungal therapy [6,8,9]. Occasionally refractory or a typical case not responding to standard therapy is treated with potassium iodide or Laser vaporization therapy. Despite all these measures the outcome is variable with almost 50% of cases improve while on treatment, 30% achieve total cure while 12% fail treatment. Those with extensive disease are most at risks of treatment failure and disease recurrence [10,11].

The case presented like deep seated mycetoma-like lesion hence wide excision with a tourniquet approach was performed to avoid seeding. Histological examination of the obtained surgically excised tissues was able to direct diagnosis towards correct assessment with the unique characteristic histological finding of granulomatous inflammation, muriform cells and Medlar bodies supporting the diagnosis. This is not exceptional since histological examinations plays pivotal role in diagnosis of rare tropical infections particularly fungal and parasitic diseases which are not common at different geographical settings [12,13].

This case report outlines the clinical presentation and management of the chronic fungal infection chromoblastomycosis at a remote health setting from endemic areas because of international travel and migration. Careful assessment with detailed history to demonstrate the occupational link, chronicity of disease is vital for correct initial assessment. For deep seated infections; radiological investigation particularly MRI are helpful towards initial assessment while histological examinations are crucial in outlying the unique pathology. Best available recommendations for deep seated

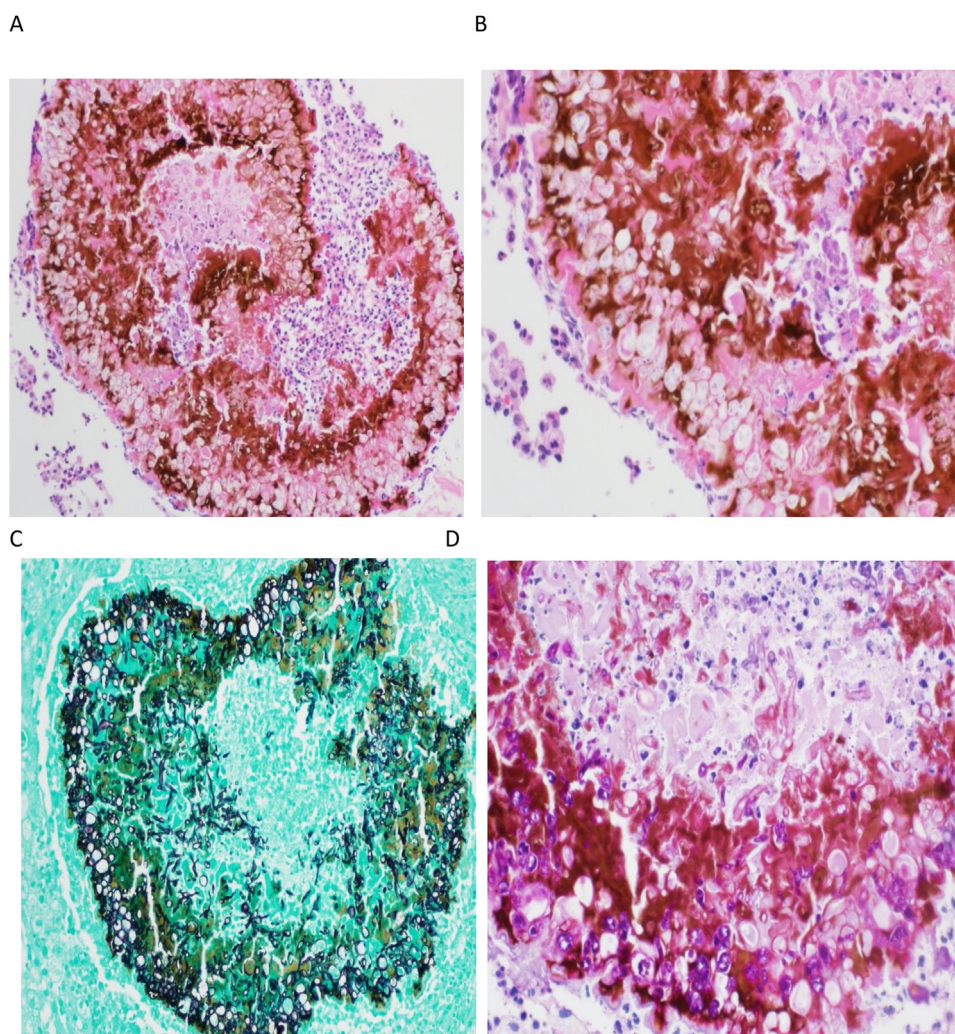


Fig. 2. Numerous brown pigmented fungal organisms amidst a zone of marked acute inflammation (H&E X 200). Rounded heavily pigmented thick-walled fungal cells (H&E X 400); C and D - Giemsa stain (X 200) and PAS stain (X 400) highlight the fungal cells.

infections are combined surgical and antifungal treatment despite the potential possibility of recurrence of the chronic disease.

Consent compliance with ethical standards

A written informed consent was obtained from the patient to include clinical presentation together with results and imaging. This was subsequently reviewed and approved by the institution ethics and research review board.

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Declaration of Competing Interest

Authors declared no competing interests in relation to current publication.

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